CHRONIC MYELOCYTIC LEUKEMIA

(Chronic Myeloid, Chronic Myelogenous, or Chronic Granulocytic Leukemia)

Clonal myeloproliferation caused by malignant transformation of a pluripotent stem cell and characterized clinically by striking overproduction of granulocytes.

CML may occur in either sex. Although CML can occur at any age, the median age is abou 45 yr; it is uncommon before 10 yr of age.

Pathology

CML is characterized by excessive production of granulocytes, primarily in the bone marrow but also in extramedullary sites (eg, spleen, liver). Although granulocyte production predominates, the neoplastic clone includes RBC, megakaryocyte, monocyte, and even some T and B cells. Normal stem cells are retained and can emerge after drug suppression of the CML clone. The bone marrow is hypercellular, but in 20 to 30% of patients, myelofibrosis develops, usually after several years. In most patients, the CML clone progresses to an accelerated phase and final blast crisis. At this time, blast cell tumor can develop in other extramedullary sites (eg, bone, CNS, lymph nodes, skin).

Symptoms and Signs

Patients are often asymptomatic early on; CML may be diagnosed during an incidental CBC. In other patients, insidious onset of nonspecific symptoms (eg, fatigue, weakness, anorexia, weight loss, fever, night sweats, a sense of abdominal fullness) may prompt evaluation. Initially, pallor, bleeding, and easy bruisability and lymphadenopathy are unusual, but moderate or occasionally extreme splenomegaly is common (60 to 70% of cases). With disease progression, splenomegaly may increase, and pallor and bleeding occur. Fever, marked lymphadenopathy, and skin involvement are ominous developments.

Laboratory Findings

In the asymptomatic patient, the WBC count is usually < 50,000/μL. In the symptomatic patient, the WBC count is usually about 200,000/μL but may reach 1,000,000/μL. The platelet count is normal or moderately increased, and the Hb is usually > 10 g/dL. On bloo smears, all stages of granulocyte differentiation are seen, although in patients with WBC counts < 50,000/μL, immature granulocytes may be uncommon. The absolute eosinophil and basophil concentrations can be strikingly increased, but the absolute lymphocyte and monocyte concentrations may be normal. A few nucleated RBCs may be present, and bloo cell morphology is normal. The bone marrow is hypercellular on aspirate and biopsy. Even at diagnosis, some patients may have some myelofibrosis. The leukocyte alkaline phosphatase score is very low.